

**2010 Hematopoietic and Lymphoid Neoplasm
Case Reportability and Coding Manual
Effective with Cases Diagnosed 1/1/2010 and after**

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Multiple Primary Rules Text Format

Note 1: Use Multiple Primary Rules M1 through M12 **before** using the Hematopoietic DB.

Note 2: The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules [M7 - M12](#).

Rule M1 Abstract as a single primary when **minimal information** is available (such as a death certificate only (DCO) case or a pathology-report-only case). *

Rule M2 Abstract as a single primary when there is a **single histology**.*

Example 1: The diagnosis is multiple myeloma (9732/3). Abstract as a single primary.

Example 2: Multiple extraosseous plasmacytomas (9734/3) are present in the oropharynx. Abstract as a single primary.

Example 3: A single histology diagnosed by the definitive diagnostic method as defined in the Hematopoietic DB; **for example**. The patient had several provisional diagnoses but the definitive diagnostic method identifies a single histology. Abstract as a single primary.

Rule M3 Abstract as a single primary when **two or more types of non-Hodgkin lymphoma** are present in the same anatomic location(s), such as one lymph node, one organ, or one tissue.*

Example: Biopsy of cervical lymph node shows **two** different non-Hodgkin lymphomas. Abstract as a single primary.

Note 1: Do **NOT** use this rule for cutaneous lymphomas.

Note 2: When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See [Appendix C](#) for information on lymph node codes and regions.

Note 3: When the disease is in a more advanced stage, both non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3 or in an organ and that organ's regional lymph nodes or in multiple organs.

- Although both non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination non-Hodgkin lymphoma, assume that all of the nodes, tissue, and/or organs are involved with the combination of non-Hodgkin lymphomas.

Note 4: Do **not** query the Hematopoietic DB in this situation.

- Rule M4** Abstract as a single primary when both **Hodgkin and non-Hodgkin** lymphoma are present in the **same anatomic location(s)**. Hodgkin and non-Hodgkin may be present in one lymph node, one organ, or one tissue.*
Example: Biopsy of cervical lymph node shows Hodgkin and non-Hodgkin lymphomas. Abstract as a single primary.
Note 1: When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See [Appendix C](#) for lymph node codes and regions.
Note 2: When the disease is in a more advanced stage, both Hodgkin and non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3 codes or in an organ and that organ's regional lymph nodes or in multiple organs.
 - Although both Hodgkin and non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination Hodgkin and non-Hodgkin lymphomas, assume that all of the nodes, tissue, and/or organs are involved with the combination of Hodgkin and non-Hodgkin lymphomas.*Note 3:* Do **not** query the Hematopoietic DB in this situation.
- Rule M5** Abstract as multiple primaries when any of the following situations are met **
 - Hodgkin lymphoma in **one node** and non-Hodgkin lymphoma in a **different node**
Note: The involved nodes may be in the same lymph node region as defined by ICD-O-3 or in different lymph node regions as defined by ICD-O-3. See [Appendix C](#).
 - Hodgkin lymphoma in **one organ** and non-Hodgkin lymphoma in a **different organ**
 - Hodgkin lymphoma in **tissue** and non-Hodgkin lymphoma in **different tissue**
Example 1: Patient is diagnosed with Hodgkin lymphoma in the cervical lymph nodes and also with non-Hodgkin lymphoma in the inguinal lymph nodes. Abstract as multiple primaries.
Example 2: Hodgkin lymphoma in thymus and non-Hodgkin lymphoma in the tonsil. Abstract as multiple primaries.
Example 3: Hodgkin lymphoma in the brain and non-Hodgkin lymphoma in the mediastinal lymph nodes. Abstract as multiple primaries.
- Rule M6** Abstract as a single primary when a **more specific** histology is diagnosed after an **NOS** when the Hematopoietic DB Multiple Primaries Calculator confirms that the NOS and the more specific histology are the same primary.
Note 1: There are no time restrictions on these diagnoses; the interval between the NOS and the more specific histology does not affect this rule stating that the two neoplasms are a single primary.
Note 2: The Hematopoietic DB will identify these histologies as a single primary.
- Rule M7** Abstract as a single primary when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days **AND** *
 - There is documentation of one positive bone marrow biopsy*Note:* When these diagnoses happen within 21 days, it is highly possible that one diagnosis was provisional and the bone marrow identified the correct diagnosis.
- Rule M8** Abstract as multiple primaries when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days **AND** **
 - There is documentation of two bone marrow examinations, one confirming the chronic neoplasm and another confirming the acute neoplasm
- Rule M9** Abstract as a single primary when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days **AND** *
 - There is no available documentation on bone marrow biopsy*Note 1:* The two diagnoses are likely the result of an ongoing diagnostic work-up. The later diagnosis is usually based on all of the test results.
Note 2: This rule applies if both neoplasms are diagnosed simultaneously (at the same time).

- Rule M10** Abstract as multiple primaries when a neoplasm is **originally diagnosed** in a **chronic** (less aggressive) phase AND **second diagnosis** of a blast or acute phase **more than 21 days** after the chronic diagnosis. **
Note 1: **This is a change from previous rules.** Use the Hematopoietic DB to determine multiple primaries when a transformation from the chronic to a blast or acute phase occurs.
Note 2: Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.
- Rule M11** Abstract the acute phase as a single primary when a neoplasm is **originally diagnosed** in the blast or acute phase and **reverts** to a less aggressive/chronic phase and there is **no confirmation** available that the patient has been treated.
Note 1: When these diagnoses happen **within 21 days**, it is highly possible that the first diagnosis of acute disease was a provisional diagnosis.
Note 2: When the subsequent diagnosis occurs more than 21 days after the original diagnosis of acute disease it is important to follow-back to obtain information on treatment or a subsequent bone marrow biopsy that negates the diagnosis of acute disease.
- Rule M12** Abstract as multiple primaries when a neoplasm is **originally diagnosed** in the blast or acute phase and **reverts** to a less aggressive/chronic phase **after treatment.** **
Note 1: Only abstract as a multiple primary when the patient has been treated for the acute disease.
Note 2: **This is a change from previous rules.** Use the Hematopoietic DB to determine multiple primaries when a transformation from the blast or acute phase to a chronic phase occurs.
Note 3: Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.
- Rule M13** Use the Hematopoietic DB to determine the number of primaries for all cases that do **not** meet the criteria of M1-M12.

*Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

**Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

Primary Site and Histology Coding Rules Text Format

Note 1: Use the Primary Site and Histology Rules **before** using the Hematopoietic DB

Note 2: The primary site and histology coding rules are divided into nine modules. Each module covers a group of **related** hematopoietic or lymphoid **neoplasms**. However, a specific histology may be covered in more than one module.

Note 3: The modules are **not hierarchical**, but the **rules** within each module **are in hierarchical** order. Apply the rules **within each module** in order. **Stop** at the first rule that applies.

Note 4: Apply rules in Module 1 first. Then go to the **first module** that **applies** to the case you are abstracting. If the situation in your case is not covered in that module **continue** on as **directed** after the last rule in the module.

Module 1: General Instructions PH1-PH3

All hematopoietic and lymphoid neoplasms (9590/3-9992/3)

Rule PH1 Code the primary site using information from scans, documentation in the medical record, the pathology report, and from the Hematopoietic DB.

Note: For hematopoietic neoplasms the pathology report is not the automatic default standard for determining the primary site. The standard for determining primary site differs depending upon the specific histology.

Rule PH2 Code the histology diagnosed by the definitive diagnostic method(s) (see Hematopoietic DB). The definitive diagnostic method can be a clinical diagnosis, genetic test, immunophenotyping, cytology, or pathology. When a pathology report is the definitive diagnostic method, code the histology from the final diagnosis, comment on the final diagnosis, addenda to the final diagnosis, or CAP protocol.

Rule PH3 Code the primary site and histology using the medical practitioner's statement on the medical record or death certificate when none of the tests or reports defined as a definitive diagnostic method is available.

Go to the appropriate Module 2-8.

When modules 2-8 do not apply to the case being abstracted, go to [Module 9](#).

Module 2: Plasma Cell Neoplasms PH4-PH8

Solitary plasmacytoma of bone 9731/3
Plasma cell myeloma/multiple myeloma 9732/3
Extraosseous plasmacytoma 9734/3

Rule PH4 Code the primary site to the site of origin, (lymph node region(s), tissue, or organ) and code the histology extramedullary plasmacytoma (9734/3) when any of the following occur in a site other than bone

- **Plasmacytoma**
- **Extraosseous (extramedullary) plasmacytoma**
- **Solitary plasmacytoma**
- **Multiple plasmacytomas**
- **Multiple extraosseous (extramedullary) plasmacytomas**

Note 1: Extramedullary and extraosseous mean “not occurring in bone.”

Note 2: 80% of extramedullary plasmacytomas occur in the upper respiratory tract (oropharynx, nasopharynx, sinuses, and larynx) although they may occur in numerous other sites including the GI tract, lymph nodes, bladder, CNS, breast, thyroid, testis, parotid, and skin.

Note 3: Do **not** code to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

Example 1: Pathology reports a solitary plasmacytoma wrapped around L4 vertebrae, no invasion of vertebrae. Code the primary site as soft tissue (C496) and the histology 9734/3.

Example 2: Scan shows two plasmacytomas in the nasopharyngeal wall. Biopsy confirms plasmacytoma. Code the primary site nasopharynx (C119) and the histology 9734/3.

Rule PH5 Code the primary site to the **specific bone (C400-C419)** where the plasmacytoma originated and code the histology solitary plasmacytoma of bone (9731/3) when the diagnosis is

- **Plasma cell neoplasm**
- **Solitary plasmacytoma**
- **Solitary plasmacytoma of bone**
- **Solitary medullary plasmacytoma**
- **Multiple plasmacytomas**
- **Multiple plasmacytomas of bone**
- **Multiple medullary plasmacytomas**

Note 1: The most common sites are bones with active bone marrow hematopoiesis; in order of frequency these include vertebrae, ribs, skull, pelvis, femur, clavicle, and scapula.

Note 2: Do **not** code primary site to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

Rule PH6 Code the primary site **unknown (C809)** and histology solitary plasmacytoma of bone (9731/3) when the only information is that the patient had a **plasmacytoma** or a **solitary plasmacytoma**.

Example: Death-certificate-only case with underlying cause of death listed as plasmacytoma.

Rule PH7 Code the primary site **bone marrow (C421)** and the histology plasma cell myeloma/**multiple myeloma (9732/3)** when the **clinical** diagnosis is plasma cell myeloma/multiple myeloma and there is no documentation of bone marrow biopsy or the results of the bone marrow biopsy are unknown or unavailable.

Example: Death-certificate-only case with underlying cause of death listed as multiple myeloma.

Note: A clinical diagnosis of multiple myeloma may be based on amyloidosis with associated renal impairment, anemia, and/or hypercalcemia supported by radiologic evidence of multiple lytic bone lesions.

Rule PH8 Code the primary site **bone marrow (C421)** and the histology plasma cell myeloma/**multiple myeloma (9732/3)** when the diagnosis is **smoldering myeloma, indolent myeloma, evolving myeloma, plasma cell myeloma, or multiple myeloma.**

Note 1: When the proportion of plasma cells in the bone marrow is 10% or greater, the diagnosis is multiple myeloma.

Note 2: A medical record may have multiple bone marrow biopsies. If any one of the biopsies is positive for multiple myeloma, code the histology to multiple myeloma (9732/3) and the primary site to bone marrow (C421).

Example: Bone marrow biopsies: Biopsy 1: Negative. Biopsy 2: Multiple myeloma with bone marrow showing 18% plasma cells. Code the primary site bone marrow (C421) and the histology 9732/3.

When this module does not apply to the case being abstracted, go to [Module 8](#).

Module 3: Lymphoma/Leukemia (Specific neoplasms that can manifest as either leukemia or lymphoma) PH9-PH12

Blastic plasmacytoid dendritic cell neoplasm, NOS 9727/3

Burkitt cell leukemia 9826/3

Burkitt lymphoma, NOS 9687/3

Precursor B-cell lymphoblastic leukemia/lymphoma 9836/3

Precursor B-cell lymphoblastic lymphoma, NOS 9728/3

Precursor T-cell lymphoblastic lymphoma, NOS 9729/3

Small B lymphocytic lymphoma 9670/3

T lymphoblastic leukemia/lymphoma 9670/3

Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma

Note 2: Commonly lymphoma originates in lymph node region(s), tissue, or organ(s) although it will metastasize to the bone marrow when the disease is stage IV or disseminated

Note 3: Commonly leukemia originates in the bone marrow

Rule PH9 Code the primary site **bone marrow (C421)** and code the histology B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (**BCCLL/CLL**) (**9823/3**) when the diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (**BCCLL/SLL**) **AND peripheral blood is involved** (the **bone marrow** may also be involved).

Note 1: Peripheral blood involvement requires repeated CBCs with absolute lymphocyte count >5000 on repeated measures or flow cytometry that documents a clonal B-cell population in the bone marrow.

Note 2: Leukemic BCCLL will always have peripheral blood involvement. The bone marrow may or may not be involved. In later stages of the disease there may be involvement of lymph nodes, liver and spleen.

Note 3: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH10 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and code the histology **small B lymphocytic lymphoma (9670/3)** when the diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **AND** you **cannot verify** that the disease originated in the bone marrow.

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules [1](#) and [7](#) for more information on coding primary site for lymphoma.

Note 2: See [Appendix C](#) for help in identifying lymph node regions and codes.

Note 3: In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow.

Note 4: Small lymphocytic lymphoma is characterized by negative peripheral blood involvement (an absolute lymphocyte count ≤ 5000 on repeated CBCs).

Rule PH11 Code the primary site **bone marrow (C421)** and the respective histology from the list below when the diagnosis is Burkitt **lymphoma/leukemia**, precursor cell lymphoblastic lymphoma/leukemia, precursor B-cell lymphoblastic leukemia/lymphoma, or precursor T-cell lymphoblastic leukemia/lymphoma **AND** the **only** involvement is **bone marrow**.

- Burkitt cell leukemia (9826/3)
- Precursor cell lymphoblastic leukemia, NOS (9835/3)
- Precursor B-cell lymphoblastic leukemia/lymphoma (9836/3)
- T lymphoblastic leukemia/lymphoma (9837/3)

Note 1: Leukemia most commonly originates in the bone marrow. When only the bone marrow is involved, code as leukemia.

Note 2: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH12 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology to the respective histology from the list below when the diagnosis is Burkitt **lymphoma/leukemia**, precursor cell lymphoblastic lymphoma/leukemia, precursor B-cell lymphoblastic leukemia/lymphoma, or precursor T-cell lymphoblastic leukemia/lymphoma **AND** there is involvement of lymph node region(s), tissue, or organ(s).

- Burkitt lymphoma, NOS (9687/3)
- Blastic plasmacytoid dendritic cell neoplasm (previously called precursor cell lymphoblastic lymphoma, NOS) (9727/3)
- Precursor B-cell lymphoblastic lymphoma, NOS (9728/3)
- Precursor T-cell lymphoblastic lymphoma, NOS (9729/3)

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules [1](#) and [7](#) for more information on coding primary site for lymphoma.

Note 2: See [Appendix C](#) for help in identifying lymph node regions, and codes.

Note 3: In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow.

When this module does not apply to the case being abstracted, go to [Module 8](#).

**Module 4: Preleukemia, Smoldering leukemia and Myelodysplastic syndrome 9989/3
PH13**

Rule PH13 Code the primary site **bone marrow (C421)** and the histology **myelodysplastic syndrome (9989/3)** when the diagnosis is **preleukemia, smoldering leukemia, or myelodysplastic syndrome.**

When this module does not apply to the case being abstracted, go to [Module 8](#).

Module 5: Myeloid Neoplasms PH14-PH15

**Acute myeloid leukemia, NOS 9861/3
Myeloid sarcoma 9930/3**

Rule PH14 Code the primary site **bone marrow (C421)** and code the histology **9861/3** when the diagnosis is **myeloid neoplasm** or **acute myeloid leukemia, NOS AND** the involvement is **limited to bone marrow.**

Note: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH15 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ and the histology to **myeloid sarcoma (9930/3)** when the diagnosis is **myeloid neoplasm** or **myeloid sarcoma AND** the neoplasm originates in a site **other than bone marrow.**

Note 1: Most common sites are skin, lymph node(s), GI tract, bone, soft tissue, and testis. This neoplasm however, can occur in almost every site of the body other than bone. Myeloid sarcoma does **not** originate in bone marrow.

Note 2: See [Appendix C](#) for help in identifying lymph node names, chains, and codes.

For rules on coding primary site for lymphomas go to [Modules 1 and 7](#).

When this module does not apply to the case being abstracted, go to [Module 8](#).

Module 6: Coding Primary Site and Histology for Specified Lymphoma PH16-PH24

Composite lymphoma 9596/3
Diffuse large B-cell lymphoma 9680/3
Follicle cell lymphoma 9597/3
Follicular lymphoma 9690/3
Follicular lymphoma, grade 1 9695/3
Follicular lymphoma, grade 2 9691/3
Follicular lymphoma, grade 3A, 3B 9698/3
Lymphoplasmacytic lymphoma 9671/3
Waldenstrom macroglobulinemia 9761/3

Rule PH16 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and code the histology diffuse large B-cell lymphoma (DLBCL) (9680/3) when **DLBCL** (9680/3) and **follicular lymphoma** (9690/3) are present **in the same lymph node(s), tissue, or organ**

Note 1: The original pathology may identify only DLBCL although both DLBCL and follicular lymphoma are present. The DLBCL is much more aggressive than the follicular lymphoma and often masks the follicular lymphoma during the initial work-up. Because it is more aggressive, the DLBCL will respond more rapidly to treatment so the post-treatment biopsies may show a combination of DLBCL and follicular lymphoma or the post-treatment biopsy may be positive for only follicular lymphoma. The follicular lymphoma was present from the beginning but was hidden. Do not change the histology; it should remain 9680/3

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules [1](#) and [7](#) for more information on coding primary site for lymphoma.

Note 3: See [Appendix C](#) for help in identifying lymph node names, chains, regions, and codes.

Note 4: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.

Rule PH17 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology to follicular when the lymphoma is described as diffuse follicular or follicular, diffuse

Example 1: Diffuse follicular lymphoma, grade 1. Code follicular lymphoma, grade 1 (9695/3)

Example 2: Follicular lymphoma, diffuse, grade 2. Code follicular lymphoma grade 2 (9691/3).

Example 3: Grade 3 follicular lymphoma, diffuse. Code follicular lymphoma, grade 3 (9698/3).

Example 4: Follicular lymphoma, diffuse. Code follicular lymphoma, NOS (9690/3).

Rule PH18 Code the primary site to **skin** (C44_) and the histology to **follicle cell lymphoma** (9597/3) when there is **skin infiltration** with **follicle cell lymphoma** or **B-cell lymphoma, follicle type** and the involvement is limited to **skin**, or limited to **skin** and the **regional lymph nodes**.

Note: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do not code follicle cell lymphoma and do **not** code skin as the primary site. Dissemination to other sites or distant lymph nodes is uncommon and would occur late in the stage of the disease.

Rule PH19 Code the primary site to **skin** (C44_) and the histology to **large B-cell lymphoma** (9680/3) when there is **skin infiltration** with **large B-cell lymphoma** or **B-cell lymphoma, large cell type** and the involvement is limited to **skin**, or limited to **skin** and the **regional lymph nodes**.

Note: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.

Rule PH20 Code the primary site to **skin** (C44_) and the histology to **B-cell lymphoma, NOS** (9680/3) when there is **skin infiltration with B-cell lymphoma** and the involvement is limited to **skin**, or limited to **skin** and the **regional lymph nodes**.

Note: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.

Rule PH21 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology **composite lymphoma (9596/3)** when **both** non-Hodgkin lymphoma and Hodgkin lymphoma are **present in the same lymph node** region(s), tissue, or organ

Note 1: Use the composite lymphoma code when

- Both NHL and HL are present in one lymph node or multiple lymph nodes in one lymph node region.
- Both NHL and HL are present in multiple lymph nodes in one lymph node region or several lymph node regions as defined by ICD-O-3. i.e.: NHL and HL present in superior hilum and superior rectal lymph nodes.
 - Assume all lymph nodes are involved with both NHL and HL even when only one lymph node is biopsied.

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Module [1](#) and [7](#) for more information on coding primary site for lymphoma.

Note 3: See [Appendix C](#) for help in identifying lymph node names, chains, and codes.

Note 4: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma

Note 5: Do **not** use the composite lymphoma code 9596/3 when:

- NHL is present in one lymph node region and HL is present in another lymph node region i.e.: NHL in cervical lymph node(s) and HL in inguinal lymph node(s)
- NHL in liver and HL in intra-thoracic lymph nodes

Rule PH22 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology to the **numerically highest ICD-O-3 code** when two or more **non-Hodgkin lymphomas** are present in the same lymph node(s), tissue, or organ.

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules [1](#) and [7](#) for more information on coding primary site for lymphoma.

Note 2: See [Appendix C](#) for help in identifying lymph node names, chains, regions, and codes.

Note 3: Commonly lymphomas originate in lymph node region(s), tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.

Note 4: This rule does not apply when NHL is present in different sites. Examples are:

- Thymic extranodal marginal-zone B-cell lymphoma is present in the thymus and diffuse large B-cell lymphoma in the hilar lymph nodes.
- B-cell lymphoma is present in the intrathoracic lymph nodes and peripheral T-cell NHL in the liver.

Rule PH23 Code the primary site blood (C420) and the histology Waldenstrom macroglobulinemia (9761/3) when there is lymphoplasmacytic lymphoma in the bone marrow and IgM monoclonal gammopathy in the blood.

Rule PH24 Code the primary site to the involved bone marrow, lymph nodes, or lymphoid tissue and the histology lymphoplasmacytic lymphoma (9671/3) when the diagnosis is Waldenstrom macroglobulinemia OR lymphoplasmacytic lymphoma and Waldenstrom macroglobulinemia AND the bone marrow, lymph nodes OR lymphoid tissue are involved.

For additional rules on coding primary site for lymphomas go to Module [1](#) and [7](#).

When this module does not apply to the case being abstracted, go to [Module 8](#).

**Module 7: Primary Site Rules for Lymphomas Only 9590/3-9729/3
PH25-PH37**

- Rule PH25** Code the primary site to the specific lymph node region when only **one lymph node or one lymph node region** is involved.
- Rule PH26** Code the primary site mediastinal lymph nodes (C771) when the site of lymphoma is described **only** as a **mediastinal mass**.
- Rule PH27** Code the primary site intra-abdominal lymph nodes (C772) when the site of lymphoma is described **only** as a **retroperitoneal mass** or as a **mesenteric mass**.
- Rule PH28** Code the primary site inguinal lymph nodes (C774) when the site of lymphoma is described only as an inguinal mass.
- Rule PH29** Code the primary site to the specific lymph node region when **multiple** lymph node **chains** within the **same region** (as defined by ICD-O-3) are involved.
Example 1: Code intra-abdominal lymph nodes (C772) when there is involvement of hepatic (C772) and para-aortic lymph node chains (C772).
Example 2: Code lymph nodes of head, face and neck (C770) when there is involvement of cervical (C770) and mandibular (C770) lymph node chains.
Example 3: Code mediastinal lymph nodes (C771) when bilateral mediastinal lymph nodes are involved.
- Rule PH30** Code the primary site as multiple lymph node regions, NOS when multiple lymph node regions (C778) as defined by ICD-O-3 are involved and it is **not possible to identify the lymph node region where the lymphoma originated**.
Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules [1](#) and [7](#) for more information on coding primary site for lymphoma.
Note 2: See [Appendix C](#) for help in identifying lymph node names, chains, regions, and codes.
Example 1: Cervical and intrathoracic lymph nodes involved with B-cell lymphoma. Code the primary site to lymph nodes of multiple regions (C778).
Example 2: CT scans showed involvement of the cervical lymph nodes (C770) and the mediastinal lymph nodes (C771). No additional involvement was identified during the work-up. Biopsy of a cervical lymph node confirmed lymphoma. Code the primary site to lymph nodes of multiple regions (C778).
- Rule PH31** Code the primary site to lymph nodes, NOS (C779) when lymph node(s) are involved but **no primary site/particular lymph node region** is identified.
- Rule PH32** Code the primary site to bone marrow (C421) when lymphoma is **present only in the bone marrow**.
Note: All available physical exams, scans, and other work-up must be negative for lymph node, tissue, or organ involvement.
- Rule PH33** Code the primary site to the specific organ when **lymphoma** is present only in an **organ**.
Note: Includes lymphomas that are primary in the spleen. Although these lymphomas are rare, if the physician states that spleen is the organ of origin, code the primary site spleen (C422).
Example: Pathology from stomach resection shows lymphoma. No other pathologic or clinical disease identified. Code the primary site to stomach, NOS (C169).
- Rule PH34** Code the primary site to the lymph node region as defined by ICD-O-3 when there is **proof of extension from the regional lymph nodes** into the organ.
Example: Patient presents with abdominal adenopathy. Surgical exploration documents direct invasion of the stomach from the regional lymph nodes. Code abdominal lymph nodes (C772).

Rule PH35 Code the primary site to the organ when lymphoma is present in an **organ** and that organ's **regional lymph nodes**.

Note: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes.

Example 1: Lymphoma is present in the kidney and peri-renal lymph nodes. Code the primary site to kidney (C649).

Example 2: Lymphoma is present in the stomach and the gastric lymph nodes. Code the primary site to stomach, NOS (C169).

Example 3: Lymphoma is present in the spleen and the splenic lymph nodes. Code the primary site spleen (C422).

Rule PH36 Code the primary site to lymph nodes, NOS (C779) when **lymphoma** is present in an **organ(s)** and **lymph nodes** that are not **regional** for that organ and the **origin cannot be determined** even after consulting the physician.

Note 1: Lymphoma can spread from organs to **regional** lymph nodes, but does **not** spread from the organ directly to **distant** lymph nodes

Example: The patient has positive mediastinal (C771) and cervical (C770) lymph nodes and involvement of the stomach (C169). No further information is available. Code to lymph node, NOS (C779).

Note 2: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes.

Note 3: See [Appendix C](#) for help in identifying lymph node names, chains, regions, and codes.

Rule PH37 Code primary site to unknown primary site (C809) only when there is no evidence of lymphoma in lymph nodes AND the physician documents in the medical record that he/she suspects that the lymphoma originates in an organ(s). See ICD-O-3 Rule D.

When this module does not apply to the case being abstracted, go to [Module 8](#).

**Module 8: Histology Rules Only: All hematopoietic and lymphoid neoplasms 9590/3-9992/3
PH38 - PH39**

Rule PH38 Code the non-specific (NOS) histology when the diagnosis is

- **One non-specific histology AND**
- **Two or more specific histologies AND**
- The hematopoietic DB multiple primaries calculator documents the specific histologies and NOS are the **same primary AND**
- No further information is available

Note 1: Use Appendix E: Histology "NOS" Tables to identify the NOS histologies.

Note 2: Use the Hematopoietic DB multiple primaries calculator to confirm that the NOS and specific histologies are the same primary.

Example: The diagnosis is myeloproliferative disorder, NOS (9960/3), polycythemia vera (9950/3), essential thrombocythemia (9962/3). The Hematopoietic DB multiple primaries calculator shows myeloproliferative disorder and polycythemia vera are the same primary. The multiple primaries calculator also shows myeloproliferative disorder and essential thrombocythemia are the same primary. Follow-back produces no additional information. Code the histology myeloproliferative disorder, NOS (9960/3).

Rule PH39 Code the specific histology when the diagnosis is

- One non-specific (NOS) histology AND
- One specific histology AND
- The Hematopoietic DB multiple primaries calculator documents the specific histology and NOS are the same primary

Note 1: Use Appendix E: Histology "NOS" Tables to identify the NOS histologies.

Note 2: Use the Hematopoietic DB multiple primaries calculator to confirm that the NOS and specific histology are the same primary.

When this module does not apply to the case being abstracted, go to [Module 9](#).

**Module 9: Default Rules: All hematopoietic and lymphoid neoplasms 9590/3-9992/3
PH40-PH41**

Rule PH40 Use the **Hematopoietic DB** to determine the primary site and histology when rules PH1-PH39 do **not** apply.

Rule PH41 When the histology code cannot be determined using the Hematopoietic DB, code the histology with **the numerically higher** ICD-O-3 code.

This is the end of the rules for coding primary site and histology.

Grade of Tumor Rules Text Format

Note 1: Use the Grade of Tumor Rules (G1-G11) to assign the correct code in the grade field.

Note 2: Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade.

Rule G1 Code cell type not determined, not stated, not applicable (code 9) for the following myeloproliferative neoplasms, myeloproliferative/myelodysplastic syndromes, myelodysplastic syndrome, histiocytic and dendritic cell neoplasms

9740/3	9946/3
9741/3	9950/3
9742/3	9961/3
9751/3	9962/3
9755/3	9963/3
9757/3	9964/3
9758/3	9975/3
9759/3	9980/3
9801/3	9982/3
9805/3	9982/3
9806/3	9983/3
9807/3	9985/3
9808/3	9986/3
9809/3	9989/3
9875/3	9991/3
9876/3	9992/3
9945/3	

Note 1: These neoplasms do not have a specific codable phenotype

Note 2: See Tables [B1](#), [B3](#), [B4](#), and [B11](#) in [Appendix B](#) for neoplasm terms and codes.

Rule G2 Use statements from **any** part of the medical record including, but not limited to

- Pathology report **OR**
- History and physical **OR**
- Consultation **OR**
- Final diagnosis **OR**
- Face sheet

Rule G3 Use codes 5, 6, 7, 8, and/or 9 **only** -- even if there is a statement giving the cell type in the medical record

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- Note 1:** Do **not** code descriptions “low grade,” intermediate grade,” or “high grade” in the Tumor Grade field. These terms refer to the Working Formulation categories of lymphoma diagnosis.
- Note 2:** Do **not** code the descriptions “Grade 1,” “Grade 2,” or “Grade 3” in the Tumor Grade field. These grades represent histology types of lymphoma rather than differentiation.

Rule G4 Code T-cell (code 5) for the following neoplasms; **T-cell** is part of the neoplasm name or the neoplasm is of **T-cell origin**

9701/3
9702/3
9705/3
9708/3
9709/3
9714/3 (unless pathologist specifically designates as a B-cell)
9716/3
9717/3
9718/3
9724/3
9725/3
9726/3
9827/3
9831/3
9834/3
9837/3

- Note 1:** Record T-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention T-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.
- Note 2:** When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Rule G5 Code T-cell (code 5) when the neoplasm is identified as **T-cell, T-cell phenotype, T-precursor, Pre-T, gamma-delta-T, or null-cell T-cell**

Rule G6 Code B-cell (code 6) for the following **B-cell precursor lymphoid neoplasms** and the **mature B-cell neoplasms**

9591/3	9684/3
9596/3	9687/3
9597/3	9688/3
9670/3	9689/3
9671/3	9690/3
9673/3	9691/3
9678/3	9695/3
9679/3	9698/3
9680/3	9699/3(continued on next page)

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9712/3	9813/3
9728/3	9814/3
9731/3	9815/3
9732/3	9816/3
9734/3	9817/3
9737/3	9818/3
9738/3	9823/3
9762/3	9833/3
9811/3	9836/3
9812/3	9940/3

- Note 1:** Record B-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention B-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.
- Note 2:** When the medical record or pathology report contains one of these terms with a different phenotype (T-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.
- Note 3:** See Tables [B7](#) and [B8](#) in [Appendix B](#).

Rule G7 Code B-cell (**code 6**) when the disease is identified as **B-cell, B-cell phenotype, B-precursor, pre-B, or null-cell and B-cell.**

Rule G8 Code Null cell, non-T non-B (code 7) when the disease is described as **null cell, non-T non-B, or common cell.**

Rule G9 Code **NK-cell** (natural killer cell) (code 8) for the following neoplasms; NK-cell is a part of the **neoplasm's name**

9719/3
9948/3

- Note 1:** Record NK-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention NK-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.
- Note 2:** When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, T-cell, or null-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.
- Note 3:** See [Table B9](#) in [Appendix B](#).

Rule G10 Code Natural Killer (NK) cell (**code 8**) when the disease is described as **NK cell, natural killer cell, nasal NK/T-cell lymphoma, or null-cell and NK cell.**

Rule G11 Code cell type not determined, not stated, not applicable (code 9) when

- There is **no statement describing the cell type OR**
- The cell type is described as **combined T AND B cell OR**
- The cell type is described as **combined B AND NK cell**

Note: There is a new site-specific factor to collect combination cell types for hematopoietic or lymphatic neoplasms in the Collaborative Stage Data Collection System, Version 2.